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Poland's anomaly is an uncommon congenital aberration of the chest wall characterized by absence of the pectoralis major muscle and other nearby musculoskeletal components. In this series, a wide spectrum of thoracic deformities was associated with the Poland anomaly, ranging from segmental agenesis of the ribs, sternum, and nearby muscles, to simple aplasia of the pectoralis major muscle. Although little disability was associated with the syndrome, the patients primarily sought operative correction due to the asymmetry and their perception of adverse cosmesis. Over a ten-year period, 53 operations were

Poland's Anomaly

Natural History and Long-Term Results of Chest Wall Reconstruction in 33 Patients

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Poland's anomaly is an uncommon congenital aberration of the chest wall characterized by absence of the pectoralis major muscle and other nearby musculoskeletal components. In this series, a wide spectrum of thoracic deformities was associated with the Poland anomaly, ranging from segmental agenesis of the ribs, sternum, and nearby muscles, to simple aplasia of the pectoralis major muscle. Although little disability was associated with the syndrome, the patients primarily sought operative correction due to the asymmetry and their perception of adverse cosmesis. Over a 10-year period, 53 operations were performed on 27 individuals with the goal of correcting the abnormal contour of the chest wall. The most successful reconstructions involved the use of the latissimus dorsi muscle, which was detached and transferred to the anterior chest wall while preserving the neurovascular pedicle. In women, this was accompanied by insertion of a mammary prosthesis. Reconstruction of the so-called "herniation" of the lung with rib grafts or alloplastic materials was found to be unnecessary, and the use of custom-made chest wall prostheses is not recommended, since four of five of these devices had to be removed due to migration, erosion of local tissues, and adverse cosmesis.

POLAND'S ANOMALY, described almost 150 years ago by Alfred Poland, is an uncommon congenital aberration of the chest wall.^{1,2} Confined unilaterally to the thorax and upper extremity, the deformity has come to include a constellation of hypoplastic components, the most consistent of which is a partial or total absence of the pectoralis major muscle. In its severest configuration, there may be partial agenesis of the ribs and sternum; scoliosis; brachysyndactyly; mammary aplasia; and absence of the latissimus dorsi, serratus anterior, and other nearby structures.³⁻⁷ Renal hypoplasia,

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certain leukemias, and the Mobius syndrome have also been described in association with the chest wall defects.⁸ An engaging account of the disorder is found in Ravitch's treatise on congenital deformities of the chest wall.⁹

The reconstruction of the defect has changed considerably over the past decade, and although newer techniques have been described, even the largest series includes relatively few patients.¹⁰⁻¹³ Most of these reports describe a singular mode of reconstruction without analyzing the natural history of the syndrome, other surgical options, or long-term results of those options. The purpose of this paper is to relate our evolving experience with Poland's anomaly in 33 patients over the last 10 years.

Materials and Methods

Thirty-three individuals who were born with unilateral absence of all or a major portion of the pectoralis major muscle were studied. All patients were evaluated by the authors over a 10-year period from 1977-1987, with 28 seen at Walter Reed Army Medical Center and 5 at Eisenhower Army Medical Center. Careful physical examinations, muscle testing, and standard photography were performed on all patients to assess and document the extent of the anomaly. Follow-up ranged from 1-27 years from their initial evaluation (several patients were seen many years ago at Walter Reed and were called in for a follow-up examination). There were 25 female and 8 male patients, ranging from 2 months to 34 years of age at the time of the initial evaluation (average = 18.9 years). All 33 had unilateral absence of the pectoralis major muscle, with 6 patients having the full-blown chest wall syndrome,

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including absence of both upper and lower portions of the muscle as well as absence of the anterior portions of ribs 3 through 5 on the same side. This latter group also had a variety of other unilateral defects including shortening and cartilaginous deformities of the sternum (sternal bar formation), hypoplasia of the clavicle and scapula, xiphoid bifidity, and pseudoherniation of the lung (Fig. 1). The other 25 patients had absence of the large sternocostal portion of the muscle with preservation of a subclavicular strip. All 33 patients had breast abnormalities with concomitant nipple-areolar findings, ranging from athelia to cephalad displacement and hypoplasia. One male patient had simple syndactyly with brachydactyly involving the index, middle, and ring fingers. Another male patient exhibited ectrodactyly (severe shortening and webbing), also involving primarily the central three rays. Seven patients had noticeable shortening of the skeletal components of the upper extremity. In addition to the



FIG. 1. This 33-year-old man was seen 27 years after his initial evaluation in our clinic. He exhibits the full-blown deformity as described and illustrated by Froriep in the original article.² Although not previously noted, the rectus abdominis muscle often inserts abnormally high, over a "sternal bar" of cartilage that is always adjacent to the lung herniation.

TABLE 1. Reconstructive Chest Wall Procedures in 27 Patients With Poland's Anomaly

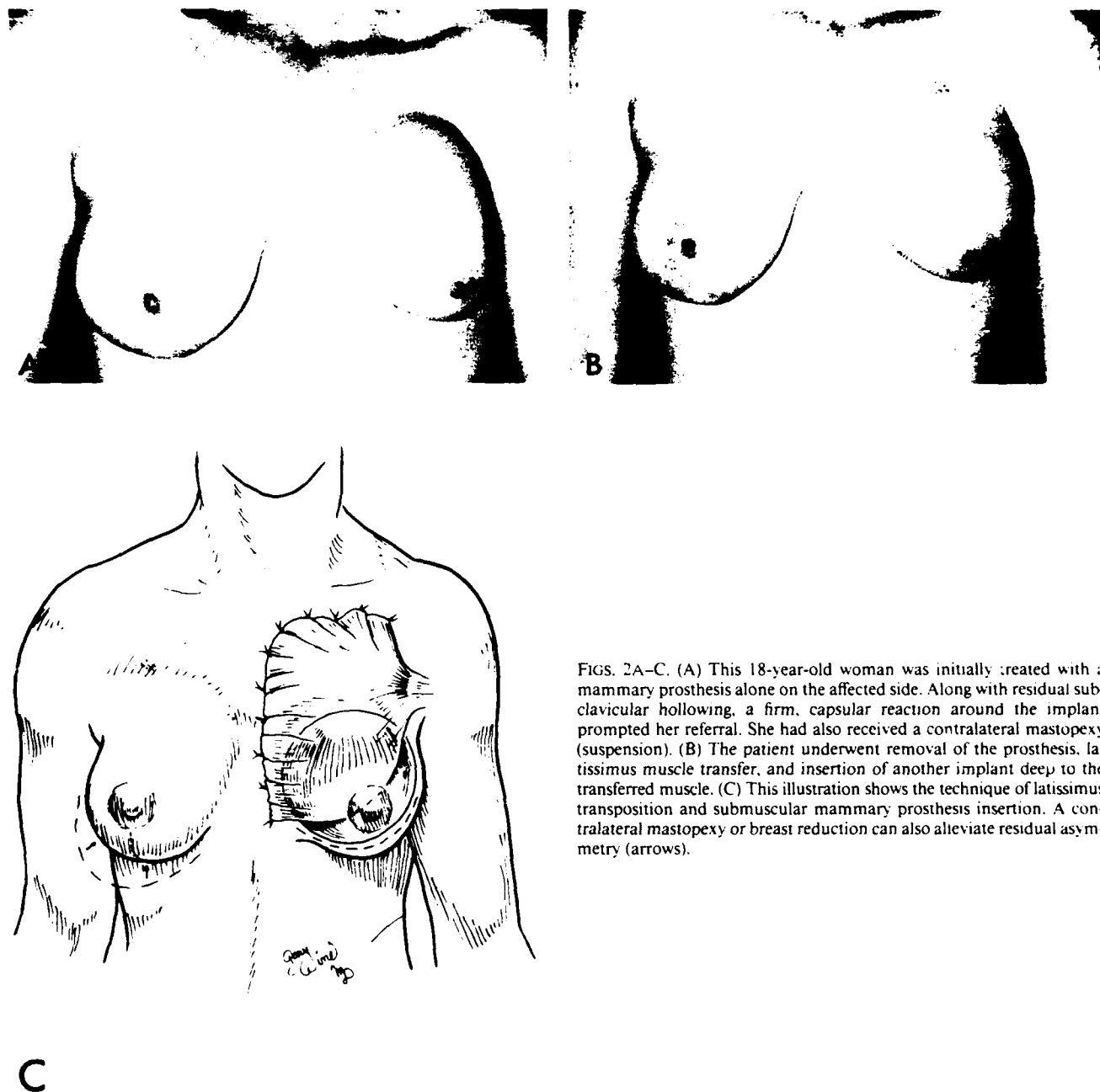
Initial Operation	Revision	Contralateral Procedures	Nipple/Areolar Procedures
I. Women			
Mammary Prosthesis			
12	5	6	2
	1		
	Latissimus transfer		
Latissimus transfer plus mammary prosthesis			
4		4	
Chest wall prosthesis			
4	4	3	
Rib reconstruction; mammary prosthesis			
1			
II. Men			
Latissimus transfer			
5	1		1
	(chest wall prosthesis; later removed)		
Sternal-rib reconstruction; latissimus transfer			
1			1

pectoralis deficiency, five patients had partial or complete absence of the ipsilateral latissimus dorsi muscle, and nine patients had absence of the serratus anterior muscle, with winged scapula. Four individuals exhibited a moderate degree of scoliosis that was concave in the direction of the deformity. The defect was located on the left side in 12 patients and on the right side in 21. In the male patients, there were five right-sided defects and three on the left. In the female patients, there were 16 defects on the right side and 9 on the left.

Results

Natural History

There was little disability associated with the disorder, even the severe form, from infancy through adulthood. However, one female patient, who also had absence of the latissimus dorsi muscle, reported difficulty in performing cartwheels and other gymnastic maneuvers due to lack of shoulder strength. Most of the male patients participated in athletics. However, with the severe form, there was weakness in performing rope-climbing and pull-ups. One man sustained an ipsilateral humeral fracture on two separate occasions while landing after hang-gliding. The hand deformities caused little disability except for the subjective complaint of decreased power grip. This parameter was not measured, however.



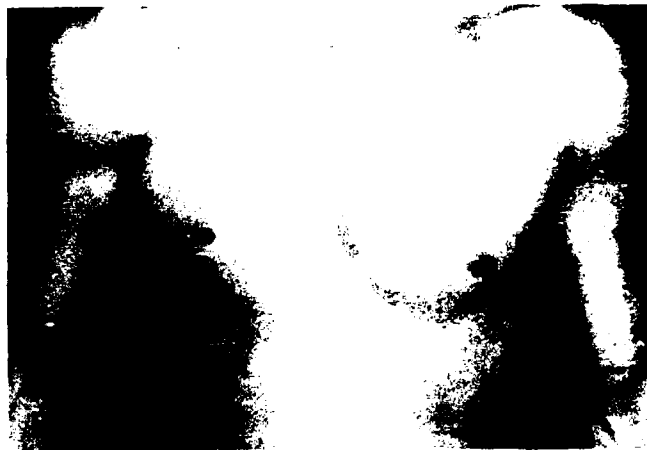
FIGS. 2A-C. (A) This 18-year-old woman was initially treated with a mammary prosthesis alone on the affected side. Along with residual subclavicular hollowing, a firm, capsular reaction around the implant prompted her referral. She had also received a contralateral mastopexy (suspension). (B) The patient underwent removal of the prosthesis, latissimus muscle transfer, and insertion of another implant deep to the transferred muscle. (C) This illustration shows the technique of latissimus transposition and submuscular mammary prosthesis insertion. A contralateral mastopexy or breast reduction can also alleviate residual asymmetry (arrows).

All patients, other than small children, complained of embarrassment due to a self-perceived cosmetic deformity. In men, this perception consisted of asymmetry and lack of soft tissue "fill" of the upper (subclavicular and parasternal) chest wall. The relative exposure of the ribs in this area, often more noticeable due to hypoplastic serratus slips, also caused concern. This was accentuated even more by a well-developed contralateral pectoralis major muscle. In women, the perception was one of extreme asymmetry due to a similar absence of subclavicular and

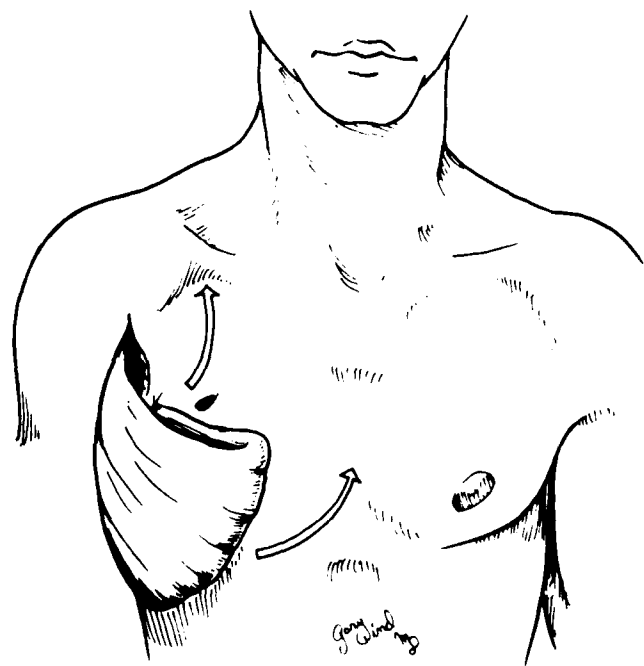
upper chest wall "fill." This was compounded by the obvious absence or hypoplasia of the ipsilateral breast.

Reconstructive Operations

Fifty-five chest wall operations in 27 patients were performed with the goal of restoring symmetry (average 2.04 procedures per patient) (Table 1). Five (three women and two men) declined treatment and one child (female) continues to be followed. In the female group that underwent



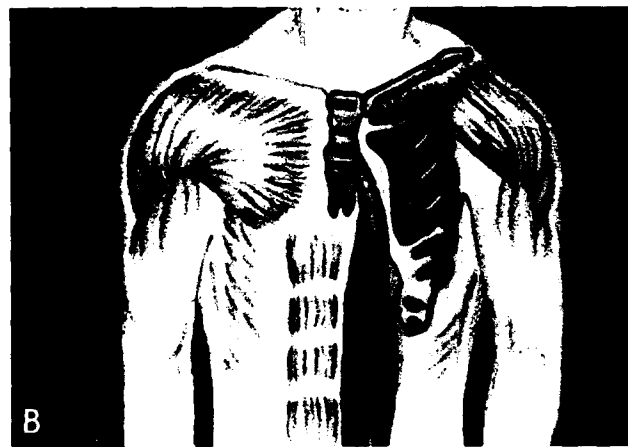
FIGS. 3A-C. (A) 17-year-old boy with the mild form of the anomaly, showing absence of the large sternocostal head of the pectoralis major muscle and relative prominence of the ribs. (B) The same patient is seen after latissimus dorsi muscle transposition and anterior transfer of the humeral insertion. (C) This illustration shows the technique of the muscle transfer in males, with folding of the distal muscle to augment parasternal bulk.



C

reconstruction ($n = 21$), the initial operation was the insertion of a mammary prosthesis ($n = 12$); a soft, custom-made chest wall prosthesis ($n = 4$); or a tissue expander followed by a latissimus dorsi muscle transfer and mammary prosthesis ($n = 1$). Rib reconstruction and later mammary prosthesis ($n = 1$) and latissimus transfer with mammary prosthesis ($n = 3$) were performed on the remainder. Eight of those who received an initial mammary prosthesis desired no further chest wall reconstruction, although two had a later areolar restoration. Of the other four patients, all underwent secondary revisions, with three receiving a larger mammary prosthesis and one re-

ceiving a smaller one. One of these also underwent a later latissimus transfer plus sublatissimus insertion of a mammary prosthesis due to persistent subclavicular hollowing and capsular contracture about the original implant (Figs. 2A-C). Of the four patients who received custom chest wall devices, three underwent removal of the implants due to migration and adverse cosmesis, despite the presence of velour backings and tabs that had been sutured to the clavicular periosteum. The remaining individual was lost to follow-up after the first year. In each of these patients, the chest wall prosthesis was ultimately replaced with a mammary prosthesis, although in one individual



FIGS. 4A-C. (A) This 21-year-old man has the severe form of the anomaly. Note the prominent parasternal "bar," which is composed of deformed cartilage surmounted by a high-inserting rectus abdominis muscle. (B) This artist's schematic of the defect shown in the previous figure depicts the shortened, bifid sternum and adjacent "herniation" of the lung found during the operation. (C) The same patient is seen after sectioning and repositioning of the sternal components into a more natural configuration, later latissimus muscle transfer, and translocation of the nipple/areola inferiorly.

the chest wall device was repositioned twice before final removal. In those patients receiving a latissimus transfer ($n = 5$), there was no apparent advantage in delaying the sublatissimus insertion of a mammary prosthesis ($n = 2$) as opposed to insertion immediately ($n = 3$). No later revisions have been required in this entire (latissimus) group. Thirteen women also underwent contralateral operations to alleviate residual asymmetry. These consisted

of reduction mammoplasty ($n = 5$), augmentation mammoplasty ($n = 5$), mastopexy ($n = 2$), or a combination thereof ($n = 1$).

Of the men that underwent reconstruction ($n = 6$), five had a latissimus transfer as the initial procedure and one underwent sternal reconstruction followed by a later latissimus transfer (Figs. 3A-C and 4A-C). One of these patients had secondary insertion of a custom-contoured

chest wall prosthesis. As in the female group, this device had to be removed later due to migration, soft tissue erosion, and adverse cosmesis. After removal, the patient was satisfied with his appearance. The other individuals who received a latissimus transfer were also satisfied and desired no further chest wall reconstruction, although one later had the nipple-areola moved inferiorly to a more symmetric location.

Transfer of the insertion of the latissimus in order to improve the axillary fold deformity was performed in three patients (two men, one woman) after the muscle had been transposed to the anterior chest wall.

There were no deaths, infections, or other complications in this series. Latissimus transfer was associated with no measurable disability. In a separate (unpublished) study, eight patients (who had the latissimus entirely removed and used elsewhere as a free flap) were tested on a computerized muscle-testing apparatus (Cybex[®], Cybex Division of Lumex, Inc., Ronkonkoma, NY). They were able to regain any measurable loss of power within 8 weeks after the procedure as compared with preoperative values. This has correlated well with clinical experience, and it appears that other muscles about the shoulder compensate for the lost latissimus dorsi function (adduction, internal rotation, and extension of the glenohumeral joint).

Discussion

It is apparent that there is a spectrum of chest wall findings in patients with Poland's syndrome. This diversity ranges from simple absence of the sternocostal head of the pectoralis major muscle to the full-blown complex and segmental absence of all parietal components of the chest wall except skin and pleurofascial membrane. Although the operations are relatively short in duration and are not associated with measurable morbidity, the large number of procedures performed in this series reflects the complexities involved in restoring symmetry, the limited goals attainable with severe defects, and the significant investment in time and commitment made by the patients.

In the main, the chest wall aberration is cosmetic in nature, and there was little associated disability even in the most severe configuration. If there is a problem, it is usually related to an upper extremity or shoulder girdle weakness.

The goal of the reconstructive effort involves the restoration of a more natural contour to the deficient area. In women, the provision of a symmetric breast mound, natural-appearing nipple-areolar complex, and adequate subclavicular "fill" is the aim of treatment. The best cosmesis in this group was accomplished utilizing a latissimus dorsi muscle to efface the subclavicular hollowing, along with a simultaneous sublatissimus mammary prosthesis to restore the breast mound. A reasonable result also fol-

TABLE 2. *Reconstructive Alternatives in Poland's Syndrome*

	Men	Women
Mild form:	Latissimus dorsi muscle transposition	<ol style="list-style-type: none"> 1. Latissimus dorsi muscle transposition with simultaneous submuscular mammary prosthesis 2. Mammary prosthesis alone 3. Selective contralateral mammary procedure (for symmetry)
Severe form:	<ol style="list-style-type: none"> 1. Sternal reconstruction (trimming/repositioning of cartilage) 2. Later latissimus dorsi muscle transposition 	<ol style="list-style-type: none"> 1. Sternal reconstruction (trimming/repositioning of cartilage) 2. Later transposition with simultaneous submuscular mammary prosthesis

lowed the insertion of a mammary prosthesis alone in the mild form of the disorder. In the man, the restoration is more difficult since a mammary prosthesis cannot be used to augment the overlying soft tissues and further camouflage the musculoskeletal deficiency. Again, the latissimus dorsi muscle was found to be the most effective substitute for the absent pectoralis major muscle. Not only does it restore missing tissue with tissue of the same type, but since the neurovascular pedicle is preserved, it retains voluntary contraction and seems to be functionally dispensable to the upper extremity. Likewise, after transfer anteriorly, the muscle fascicles course in the same general direction as the normal pectoralis fascicles. After this is accomplished, the ribs, being covered by the transposed muscle, become much less noticeable (Table 2).

The abnormality of the anterior axillary fold remains a problem, even after a successful latissimus transposition. This is due to the unique configuration of the normal insertion of the pectoralis major and its spiralling lowermost fibers. Although detachment and anterior transfer of the latissimus insertion were accomplished in three patients, this maneuver failed to produce a completely natural axillary fold. Thorough transaxillary widening of the tunnel through which the latissimus is drawn also helped by favorably directing the fibers of the muscle toward the distal sternum. If the latissimus is also missing, the rectus abdominis may be considered for transfer. Although we have used this option for other chest wall deformities, it was purposely avoided in this series. There are cogent reasons for preserving this muscle in young, healthy subjects, not the least of which is later abdominal wall laxity or weakness.

Past techniques to "stabilize" the thorax in the severe form of the anomaly have included rib transposition or grafting, prosthetic mesh, despeciated materials, and me-

tallic sheets.¹² However, although autologous rib grafts were used in one patient early in the series, these methods were found to be unnecessary in the other individuals. The paradoxical motion of the pleurofascial membrane was not severe enough to clinically impair ventilatory mechanics in any patient, and this layer was so sturdy that no patient sustained any mishap due to weakness or absence of rib support in this area. In such defects, the badly protruding costosternal cartilage can be resected or repositioned and the abnormally inserting rectus abdominis muscle relocated inferiorly. The contour can then be improved with a latissimus transfer.

It is important to recognize that this defect commonly involves the entire hemithorax. The findings may be subtle, but many patients exhibit clavicular or humeral shortening, deficiency of the parascapular musculature, and hypoplasia of other skeletal components. This fact tends to place any potential goals into perspective.

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